Case Report

Huge Left Atrium Accompanied by Normally Functioning Prosthetic Valve

Feridoun Sabzi, MD*

Imam Ali Hospital, Kermanshah University of Medical Sciences, Kermanshah, Iran.

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Abstract

Giant left atria are defined as those measuring larger than 8 cm and are typically found in patients who have rheumatic mitral valve disease with severe regurgitation. Enlargement of the left atrium may create compression of the surrounding structures such as the esophagus, pulmonary veins, respiratory tract, lung, inferior vena cava, recurrent laryngeal nerve, and thoracic vertebrae and lead to dysphagia, respiratory dysfunction, peripheral edema, hoarse voice, or back pain. However, a huge left atrium is usually associated with rheumatic mitral valve disease but is very rare in a normally functioning prosthetic mitral valve, as was the case in our patient. A 46-year-old woman with a past medical history of mitral valve replacement and chronic atrial fibrillation was admitted to our hospital with a chief complaint of cough and shortness of breath, worsened in the last month. Physical examination showed elevated jugular venous pressure, respiratory distress, cardiac cachexia, heart failure, hepatomegaly, and severe edema in the legs. Chest radiography revealed an inconceivably huge cardiac sell-out. Transthoracic echocardiography demonstrated a huge left atrium, associated with thrombosis, and normal function of the prosthetic mitral valve. Cardiac surgery with left atrial exploration for the extraction of the huge thrombosis and De Vega annuloplasty for tricuspid regurgitation were carried out. The postoperative course was eventful due to right ventricular failure and low cardiac output syndrome; and after two days, the patient expired with multiple organ failure. Thorough literature review showed that our case was the largest left atrium (20 × 22 cm) reported thus far in adults with a normal prosthetic mitral valve function.

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Introduction

Extreme left atriomegaly was first reported as "aneurysmal dilatation of the left auricle" by Fasseas,[1] and an early autopsy report of a huge left atrium (LA) was published by Kronzon.[2] Elsewhere, a case of the massive enlargement of the LA was reported by Schwartzman.[3] The reported incidence of the huge LA in rheumatic mitral valve disease was 3 in each 1000 operations according to Piccoli.[4] Huge enlargement of the LA is usually associated with rheumatic mitral stenosis or regurgitation, left ventricular failure, chronic atrial fibrillation, and left-to-right shunts such as those occurring with patent ductus arteriosus and ventricular septal defects. Piccoli et al.[4] stated that enlargement of the LA may create compression of the surrounding structures such as the esophagus, pulmonary veins, trachea, left main bronchus,

*Corresponding Author: ***Feridoun Sabzi,*** *Professor of Cardiac Surgery, Kermanshah University of Medical Sciences, Imam Ali Hospital, Shahid Beheshti Ave., Kermanshah, Iran. Tel: +98 9181311896. Fax: +98 831 8360043. E-mail: dr_sabzi@yahoo.com.*

middle and lower lobes of the right lung, inferior vena cava, recurrent laryngeal nerve, and thoracic vertebrae, leading to dysphagia respiratory dysfunction, peripheral edema, hoarse voice, or back pain. Johnson[5] stated that paradoxical movement of the left ventricular posterobasal wall occurs in the giant LA and might exert a negative and depressive effect on the patient's hemodynamic. LA dilatation mayalso cause atrial fibrillation and thromboembolism. The giant LA is defined as an LA measuring larger than 8 cm, and it is typically found in patients who have rheumatic mitral valve disease with severe regurgitation. Our thorough literature search did not yield any reports of the giant LA (20-22 cm) in normal prosthetic valve function in adults.

Case Report

A 46-year-old woman with a past medical history of rheumatic heart disease, mitral valve replacement, and chronic atrial fibrillation was admitted to our hospital with a chief complaint of cough and shortness of breath, worsened in the last month. She was referred by a cardiologist from rural areas and, despite persistent symptoms, she had not seen a physician for many years. Physical examination showed elevated jugular venous pressure, respiratory distress, cardiac cachexia, symptoms of progressive heart failure, hepatomegaly, and severe edema in the legs. Chest radiography revealed an inconceivably huge cardiac sell-out. She had undergone the Bjork-Shiley mitral valve prosthesis replacement for severe mitral stenosis when she was 21 years old, in 1987. In the Emergency Ward, a plain chest X-ray study showed a marked cardiomegaly and nearly complete opacification of the lower, middle, and upper lung fields (Figure 1). She suffered from voice hoarseness, but there were no complaints of dysphagia or any other gastrointestinal symptoms. Blood gas analysis was normal. An electrocardiogram demonstrated severe right ventricular hypertrophy and atrial fibrillation. Transthoracic echocardiographic examinations in four chamber view (Figure 2) revealed a moderate decrease of systolic function and severe regurgitation of the tricuspid valve. This examination also unexpectedly demonstrated a massively enlarged LA with a maximum diameter of 20 cm and a transverse diameter of 21 cm with huge thrombosis, marked enlargement of the right ventricles, and severe dilatation of the pulmonary artery, superior vena cava, inferior vena cava, and pulmonary artery. There was also evidence of dilated right-side heart chambers, severe tricuspid valve regurgitation, and pulmonary artery systolic pressure of 100 mmHg. Laboratory examinations revealed abnormal renal function (creatinine > 2.5 mg/dl), abnormal liver function (alanine transferase > 300U), and iron deficiency anemia (hemoglobin = 10 mg/dl). No evidence of active rheumatic disease was documented. The ultrasound study of the abdomen showed congestive hepatomegaly with ascites.

The gigantic LA is uncommon and is defined according to the chest X-ray study appearance, in which either the LA forms the right margin of the heart shadow and approximates the right chest with a cardio-thoracic ratio greater than 60% or the LA on echocardiography has an anteroposterior diameter larger than 8 cm. A normal LA is located in the middle of the chest, is the most posterior chamber of the heart, and is not located on the left; nevertheless, when it enlarges, it moves rightward and approximates the right chest margin. In our patient, the LA measured 20 × 22 cm, representing one of the largest LAs reported in an adult. The patient's systolic pulmonary artery pressure was 100 mm Hg, which was consistent with severe pulmonary hypertension.

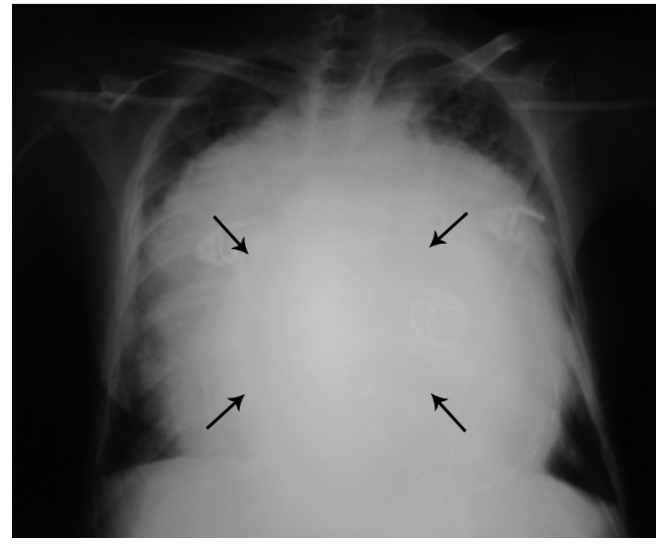

Figure 1.Chest X-Ray in posteroanterior view, showing a huge left atrium cardiomegaly (arrows)

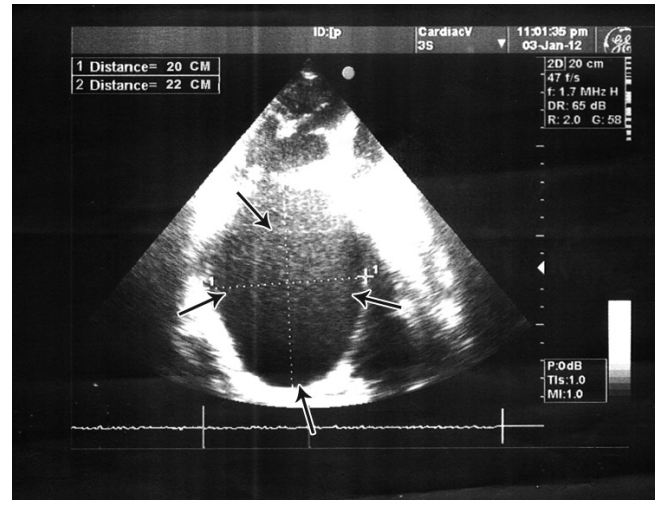

Figure 2. Echocardiography in the four-chamber view, showing a gigantic left atrium (arrows)

The chest was opened via median sternotomy. The entire heart mass was tightly adherent through relatively firm and edematous adhesions to the collapsed left and right lungs and the sternal plate. It was very difficult to find a dissection plane to reach the aorta for cannulation in consequence of severe main pulmonary artery dilatation and the enlargement of the right atrium and the right ventricular outflow tract. In order to assess the small aorta, embedded in fibrous adhesion between a huge superior vena cava and a huge main pulmonary artery, cardiopulmonary bypass (CPB) was commenced by instituting femoral artery and vein cannulation. Instituting CPB and unloading the cardiac chamber revealed the small aorta, embedded between the large superior vena cava and the main pulmonary artery. The small aorta was then released from the fibrous adhesion and prepared for aortic cross-clamping. The aorta was thereafter clamped, and cardiac arrest was induced with 1.4 liters of antegrade cold blood cardioplegia. The mitral prosthesis was fully mobile with small pannus ingrowths. A large amount of fresh and organized thrombosis was removed from the LA. Next, tricuspid valve repair without an annuloplasty ring was undertaken, and a classic De Vega procedure was performed (single Ethibond suture from the anteroseptal commissure to the posteroseptal commissure with a pledget at each end). The aorta was cross-clamped for a total of 100 minutes, and the heart function was resumed after multiple cardioversion attempts. The total CPB time was 3 hours and 20 minutes. After 20 minutes, partial bypass was resumed for a further 30 minutes because of major bleeding from the traumatic rupture of the right pulmonary artery by aortic cross-clamping.

Seven hours after the start of sternotomy, the patient was returned to the Intensive Care Unit (ICU) while she was hemodynamically unstable and had an unresponsive low cardiac output. She expired 48 hours later due to low cardiac output syndrome.

Discussion

The most common causes of the enlarged LA in adults are mitral stenosis and severe mitral regurgitation with pulmonary hypertension. A few case reports have described concomitant huge dilatation of three heart chambers in adults. In a case report on a patient with a normally functioning prosthetic mitral valve, Rahimtoola and colleagues[6] stated that the size of the left and right atria as well as that of the right ventricle increases substantially in pulmonary hypertension with non-regression of pulmonary vascular resistance and concluded that activation of the Frank-Starling mechanism occurs in both right chambers. The authors attributed these chamber enlargements to valve prosthesis-patient mismatch, underscored the problem of valve prosthesis-patient mismatch in the first generation of prosthetic valves, and asserted that LA dilatation results from the following two factors. First, the in vivo effective orifice valve area of almost all types of old prosthetic valves that can be inserted in most patients is less than that of the normal human valve. The in vivo effective prosthetic valve area is even further reduced by organized clot, pannus formation, tissue ingrowths, and endothelialization and, therefore, these valves can be considered stenotic. Second, in some patients, the problem is complicated because the size of the prosthesis that can be inserted is limited by the size of the annulus, which is small compared with the size of the patient, and also by the size of the cavity in which the prosthesis must lie. Castrillo and colleagues[7] reported a patient with a huge LA (8cm) with a normally functioning mitral valve without explaining the probable causes of the enlargement. In the Funk et al. case reports,[8] a female patient with a mitral Starr-Edwards valve prosthesis, which had been implanted 35 years previously for rheumatic mitral valve disease, was reported. Transthoracic echocardiography showed a giant LA (12cm×13cm; area of 127 cm^2) with hyperechogenic walls, mostly occupied by thrombus. She was treated conservatively; her symptoms improved mildly on diuretics, Digoxin, and anticoagulants and she was discharged 8 days after admission. Plaschkes and colleagues[9] reported the case of a 56-year-old man with an LA diameter of 17 cm as measured by echocardiography and an LA size of 18 × 20 × 17 cm according to magnetic resonance imaging. An LA diameter of 18.5 cm, reported by Ates and collogues,[10] was the largest diameter that we managed to find in the medical literature.

Castrillo and colleagues[7] demonstrated that LA enlargement is not solely due to mitral regurgitation but it is also correlated with the quality of the LA wall. Moreover, cardiac cachexia and malnutrition also predispose one to this condition. Plaschkes and colleagues[9] described partial auto transplantation for the reduction of the size of the LA and thus prevention of thromboembolism. In this technique, after cross-clamping, the superior vena cava, aorta, and pulmonary artery are detached, and the heart is easily moved upward. According to Piccolli and colleagues,[4] paradoxical movement of the left ventricular posterobasal wall occurs in the giant LA and may affect hemodynamic in a negative manner and also cause atrial fibrillation and thromboembolism. In the Lee-Roux and colleagues study,[11] radiographic evidence of asymmetrical enlargement of the LA without atrial infarction is presented. The authors stated that giant atrial enlargement is rarely symmetrical, the atrial appendage can contribute to the enlargement, and the giant atrium can be effectively trimmed. In their study, all patients with an enlarged LA (more than 6 cm), irrespective of the cause and mechanism of the enlargement, were evaluated. In the Ates and colleaguesstudy,[10] postoperative clinical and hemodynamic parameters showed a positive response to mitral valve replacement in patients with a huge LA. A direct correlation between early or late thromboembolism and the gigantic LA was not found. Inadequate control

of the anticoagulation level was a major risk factor for thromboembolism, which was encountered during the follow-up, especially in patients dwelling in rural areas. Johnson[5] found that para-annular plication, posterior wall plication, ligation of the appendix of the LA, and partial resection using auto transplantation are the possible modes of diminishing size and preventing stagnation. Goldberg and colleagues[12] described a 67-year-old woman with a history of rheumatic heart disease admitted due to dyspnea and anasarca. She had undergone the mitral valve replacement of a mechanical valve ten years earlier. Echocardiography revealed a huge LA (17 cm). The patient expired during hospitalization.

In the case of our patient, we were initially unable to define the role of atrial enlargement in determining or contributing to the patient's congestive heart failure. Be that as it may, the role of atrial enlargement in LA thrombosis formation is clear. With an apparently normal mitral prosthetic function and echocardiographic evidence of a preserved left ventricular systolic function with some diastolic restraining, Johnson and colleagues[5] hypothesized that pericardial release could increase the compliance of the left ventricle and eliminate a possible cause of limited left ventricular filling and cardiac output. We believe that in the absence of discrete constrictive pericarditis, this maneuver would not have improved our patient's cardiac output but instead would have aggravated respiratory dysfunction by causing total left lung atelectasis and worsening the patient's congestive heart failure symptoms.

The role of atrial reduction in the setting of mitral valve replacement has been widely discussed in the literature both in regard to its ventilatory effect as well as its hemodynamic effects but a consensus has not yet been reached. Parinnelo and colleagues[13] stated that LA compensation due to volume or pressure overload in mitral valve disease is, frequently, dilatation. Proposing that LA structural remodeling is independent of the atrial pressure value and effective orifice area of the mitral valve opening, and pulmonary or arterial capillary pressure values, the investigators stated that the amount of mitral regurgitation causes the enlargement of the LA. This notion supports the role of other unknown underlying factors.

Finally, severe LA enlargement is more commonly associated with mitral regurgitation than with mitral stenosis and more commonly with the rheumatic rather than with the non-rheumatic causes of mitral regurgitation. This report raises some questions vis-à-vis patients with a gigantic LA and normal prosthetic function. Indeed, does an underlying disease beget a gigantic LA or, as Parinnelo and colleagues[13] proposed, does the patient develop a giant LA in consequence of many years of mitral malfunction after valve replacement in combination with LA wall remodeling change? We believe the patient-valve mismatch in the old generation of prosthetic valves and chronic volume/pressure overload of many years are allied to the gigantic LA and that an increased trans-prosthetic mean pressure gradient and unrelieved pulmonary hypertension generate the dilated LA. In addition, the pre-existent rheumatic myocardial disease may, in all likelihood, facilitate muscular remodeling with subsequent severe atrial enlargement.

Conclusion

This case report is an attractive model for understanding the nearly unlimited LA compliance and enlargement during rheumatic valve disease. The novelty of the report appears to be the association between a huge LA (largest LA reported to date in the literature), normal prosthetic function, huge LA thrombosis associated with a gigantic superior vena cava, huge right atrium, and pulmonary artery and tricuspid valve regurgitation. The present report is also unique inasmuch as the surgeon was unable to institute cardiopulmonary bypass (CPB) via median sternotomy because the exposure of the aorta was impossible without unloading the anteriorly placed and enlarged pulmonary artery and superior vena cava.

In our patient, congestive heart failure and respiratory failure concomitant with LA thrombosis occurred with moderately reduced left ventricular dysfunction, severe right ventricular dysfunction, and a normally functioning mitral prosthesis. We were, therefore, compelled to postulate that the pathophysiology involved was created by LA enlargement in concert with pressure and volume overload and unrelieved pulmonary hypertension. In retrospect, we were faced with a patient condemned by a natural history that could not possibly be addressed by a less invasive operation. Clot removal procedure would have been the sole chance of success if the huge cardiac chambers and adhesion severity had allowed a reasonable and usual operative time.

We would believe that such tragic results make a case for the prevention of this severe complication during conventional mitral valve surgery via primary plication of the enlarged LA, even at the expense of a prolonged aortic-cross clamping time.

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